208 A review of itraconazole use in a regional paediatric CF centre

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The anti-fungal agent itraconazole has been advocated as a potential adjunct to steroid therapy in ABPA. Recent reports have highlighted the potential of itraconazole to interact with other drugs metabolised by the liver, by inhibition of CYP3A, particularly inhaled corticosteroids (ICS). In view of this and one case of iatrogenic Cushings syndrome at our clinic we reviewed our use of itraconazole.
Method: Retrospective case notes review of 106 patients, Jan 2000–06.
Results: Eleven patients (1 female; mean age 7.8 years) had been diagnosed with ABPA according to ERCF guidelines (Mastella, 2002). All 11 had prednisolone, 3/11 with ICS, 2/11 with itraconazole, 6/11 with ICS and itraconazole. Itraconazole was commenced between 0 and 20 months after diagnosis (doses range 100–400 mg daily; adequate blood levels achieved in all). Of the 8 patients on itraconazole 1 was discontinued after 2weeks;1 developed Cushings syndrome after 8 months of itraconazole and ICS therapy, subsequently making a full recovery. Six patients remain on long term itraconazole (>12 months), 5 of them combined with ICS. A short synacthen test has now been performed on 4/5 patients receiving long term itraconazole and ICS. One had an adequate response and 3 had suboptimal responses.
Discussion: Management of ABPA in Cystic Fibrosis is a challenge and the use of itraconazole may allow steroid sparing, though the evidence base to support this strategy is limited. Our data suggest that patients on itraconazole and ICS need careful monitoring and routine adrenal function testing is recommended.

209 Pregnancy in lung transplant recipients in France

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We report the French experience of 18 pregnancies in 17 women (10 with CF) who had received heart and lung (10 patients), lung (5 patients) or lung and liver transplantation (2 patients). Pregnancy occurred within 5 months to 13 yrs (mean 4.3 yrs) after lung transplantation (LT) at the mean age of 27 (18–36) yrs old.
There were 13 live births, 3 therapeutic abortions, 1 *in utero* death at 22 weeks and 1 death of the mother in the first weeks of pregnancy. Two required caesarean sections. Four babies (31%) were premature but all 13 children were well at follow-up. Low birth weight occurred in 5 newborn babies (38%). Only one child was breast fed.
Maternal follow-up after delivery ranged from 6 months to 16 years. Five (29%) maternal deaths occurred: the first one of pulmonary infection 5 months after LT as pregnancy was just discovered; the second one of pulmonary embolism after therapeutic abortion performed because of obliterative bronchiolitis (OB); the third one 1.5 year after delivery in a patient whose pregnancy had been discouraged because of pre-existing OB; the two others after a survival of 1.7 and 3 years post-partum in women whose pregnancies had occurred 1 and 8 years after LT and who developed OB after delivery.
OB developed after pregnancy in 4 of the 12 patients who are alive. For one woman, OB appeared after her second pregnancy and she was successfully retransplanted 6 years after the second delivery. The other 8 patients are well with a mean FEV_1 of 89±12% predicted.
These data show pregnancy after LT is risky, it should not be recommended before 3 years of stable condition after LT and women should be given appropriate information.